The supraspinal control of movements
Organisation of the motor system

- Sensory information
- Association cortex
- Basal ganglia
- Cerebellum
- Thalamus
- Primary motor cortex
- Brain stem
- Spinal neurones
- Motoneurones (the final "common pathway")

- Initiation of a motor activity

- PLAN
- PROGRAM
- EXECUTION
Organisation of the motor system

• Spinal cord
  – Basic patterns of posture and movement

• Brain stem
  – Postural control

• Cortex
  – Execution of target-oriented movements
## Consequences of spinal transection

<table>
<thead>
<tr>
<th></th>
<th>Acute effects</th>
<th>Chronic effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensory functions</td>
<td>Anaesthesia</td>
<td>Anaesthesia</td>
</tr>
<tr>
<td>Motor activity</td>
<td>Paralysis&lt;br&gt;Flaccidity&lt;br&gt;Areflexia</td>
<td>Paralysis&lt;br&gt;Spasticity&lt;br&gt;Hyperreflexia</td>
</tr>
<tr>
<td>Autonomic functions</td>
<td>Areflexia&lt;br&gt;No sweating&lt;br&gt;No shivering&lt;br&gt;Low blood pressure&lt;br&gt;Passive incontinence</td>
<td>Hyperreflexia (mass reflex)&lt;br&gt;Sometimes profuse sweating&lt;br&gt;No shivering&lt;br&gt;Labile blood pressure&lt;br&gt;Active incontinence</td>
</tr>
</tbody>
</table>
The clinical picture

**Spinal shock:** 3 days – 2 weeks

- Some flexor reflexes return (ankle → knee → hip) – **Babinski’ sign**

- Extensor reflexes begin to return (approx. 6 months later)

- Exaggerated extensor activity → spasticity + hyperreflexia

**Mass reflex:** stimulation of the sole evokes a highly exaggerated reflex action:
- Flexion of the legs
- Defecation and urination
- Erection

- The abdominal skin reflex and the cremaster reflex never return
Explanation of diaschisis
Regulation of posture and muscle tone
Decerebrate rigidity

Cerebral cortex

Red nucleus

Inhibitory reticular formation

Decerebration

Vestibular system

Lateral vestibular nucleus

α motoneurones

γ motoneurones of antigravity muscles
Decorticate rigidity (Wernicke-Mann posture)

Cerebral cortex

Decortication

Vestibular system

Lateral vestibular nucleus

\(\alpha\) motoneurones

\(\gamma\) motoneurones of antigravity muscles

Red nucleus

Inhibitory reticular formation
Kisagyi Purkinje-sejtek

Calbindin-specifikus jelölés

Rodaminos töltés
A cerebellum

• Cerebellum: „Head ganglion of the proprioceptive system”

• **Major functions:**
  • Maintaining balance
  • Coordination of movements – especially that of rapid movements *(corrections)*
  • Motor learning
  • Cognitive function

• **Electrical stimulation of the cerebellum does not evoke conscious sensation, and it is not followed by noteworthy movement(s)**
  
  • Although it receives sensory inputs from many sources, these do not reach conscious level

  • Although it has significant roles in the actual execution of the movements, it does not participate DIRECTLY in their completion

• It has *ipsilateral* connection with the spinal cord, BUT *contralateral* connections with the cerebral hemispheres
Functional parts of the cerebellum

• Vestibulocerebellum (the flocculo-nodular lobe)
  – Archicerebellum
  – Coordination of the trunk muscles
    • Maintaining balance
  – Coordination of the extraocular muscles

Spinocerebellum (vermis and the related cortical areas)
  – Paleocerebellum
  – Tracking and correction of movements using the proprioceptive inputs
    • Trunk and limb movements – walking (gait)

• Cerebrocerebellum (cerebellar hemispheres)
  – Neocerebellum
  – Planning and tracking of skilled movements, and cognitive function
    • Highly skilled, learned, voluntary movements
Consequences of lesions affecting the cerebrocerebellum

- Ataxia
- Dysmetria
- Intention tremor
- Dysarthria (scanning speech)
- Dysdiadochokinesis
- Adiadochokinesis
- Alteration of muscle tone
- Dyssynergia (decomposition of movements)
- Rebound phenomenon
The Marr theory

A THEORY OF CEREBELLAR CORTEX

BY DAVID MARR*

From Trinity College, Cambridge

(Received 2 December 1968)
Consequences of lesions affecting the cerebrocerebellum

• Generally: particularly strong voluntary control is required for the execution of movements – even in those situations, when it would not be necessary under physiological circumstances

• Interestingly, the chances of recovery are surprisingly – the cerebral cortex is capable of “taking over” the function of the cerebellum
Basal ganglia
Connections of the basal ganglia

• Basal ganglia receive little information from the spinal cord

• The most important input device: *neostriatum* (putamen and caudate nucleus)

• The source of the *incoming information*: cortex, hypothalamus, subthalamic nucleus, substantia nigra

• **Output channels:**
  – Down: red nucleus and reticular formation
  – Up: thalamus → precentral gyrus
Functions of the basal ganglia

• Genesis of basic movement patterns
  – The present motor programs in response to the information arriving from the association cortex

• Regulation of muscle tone and movements

• Initiation of movements based on emotional changes

• Cognitive and affective functions
Symptoms of basal ganglia disorders

• Positive (os hyperkinetic) symptoms
  – TREMOR
  – RIGIDITÁS
  – CHOREA
  – ATHETOSIS
  – BALLISMUS

• Negative (or hypokinetical) symptoms
  – HYPO- or AKINESIA
Parkinson’s disease

- **Parkinson’s trias:**
  - Akinesia
  - Rigidity
  - Tremor

- **Cause:**
  - Damage of the dopaminergic nigrostriatal pathways

- **Therapy:**
  - Administration of L-DOPA
Huntington’s chorea

- Incidence: 5-10/100 000
- Autosomal dominant
- Short arm of the 4th chromosome
The gene

- **The huntingtin gene**
  - CAG-repetition (...CAGCAGCAGCAGCAG...)

- The CAG triplet encodes glutamine $\rightarrow$ poliglutamine-(poliQ) sequence

- The healthy huntingtin contains 27 glutamines at most; if more than this $\rightarrow$ pathological huntingtin $\rightarrow$ Huntington’s disease

- The number of glutamine residues determines the onset and severity of the disease!!!
The protein (huntingtin)

- Its exact function is still unknown
- Its expression increases the chances of neural survival, whereas the presence of the mutant form increases the rate of nerve cell loss
- Anti-apoptotic effect
- Regulates the production of BDNF (brain derived neurotrophic factor)
Correlation between the number of CAG-repeats and the clinical manifestation of the disease

<table>
<thead>
<tr>
<th>Repeat</th>
<th>Classification</th>
<th>Clinical form</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;27</td>
<td>Healthy individual</td>
<td>No symptoms</td>
</tr>
<tr>
<td>27–35</td>
<td>Intermediate case</td>
<td>No symptoms</td>
</tr>
<tr>
<td>36–39</td>
<td>Reduced penetrance</td>
<td>Weak symptoms, delayed onset</td>
</tr>
<tr>
<td>&gt;39</td>
<td>Full penetrance</td>
<td>Classical form</td>
</tr>
</tbody>
</table>
Symptoms of the Huntington’s disease

Characteristic movement disorders

+ Other symptoms, including mood disorders

- Depression
- Anxiety
- Irritability
- Apathy
- Psychotic state

There is no way to prevent the onset of the disease, to alleviate the symptoms or to delay the onset.
Hemiballism

• Damage of the subthalamic nucleus on one side

• Characteristic, forceful, ballistic movements
The "extrapyramidal system"

- **Reticular formation** → lateral and medial reticulospinal tract
- **Vestibular nuclei** → vestibulospinal tract
- **Red nucleus** → rubrospinal tract
- **Tectum** → tectospinal tract
Reticulospinal tract

- Mainly ipsilateral projection
- Terminates on spinal interneurones
- Trunk and proximal muscles of the limbs are affected
- Important in maintaining the POSTURE
Vestibulospinal tract

- Mainly ipsilateral projection
- Mainly extensors (“antigravity muscles”) are affected
- Important in maintaining and controlling the posture and muscle tone
Rubrospinal tract

- Crossed pathway
- Some fibres terminate on $\alpha$ motoneurones, but it mainly targets interneurones that affect both flexors and extensors
- Lesion: it becomes difficult to perform voluntary movements, but there is no real problem with maintaining the posture
Tectospinal tract

- Originates from the tectum (superior and inferior collicles)
- Mediates the integration of auditory and visual information
- Has basic roles in ensuring proper orientation
- Proceeds to the cervical segments of the spinal cord; crossed fibres
- Terminates on interneurones that affect the movements of the head and eyes
Cortical areas involved in the motor function

- **Primary motor cortex**
  - Precentral gyrus
  - Brodmann’s 4

- **Praemotor area**
  - „Non-primary” motor cortex
  - Brodmann’s 6
    - „True” preemotor area
    - Supplementary motor area
Significance of the cortical motor areas

- **Primary motor cortex**
  - Actual performance of the motoric tasks

- **Premotor cortex**
  - The activity of this region *always precedes* that recorded from the primary motor cortex
  - Involved in the „preparation” phase of the voluntary movements
  - Isolated lesion: *apraxia* (inability to perform complex motor tasks)
The corticospinal (pyramidal) tract

• **Composition**
  – $2 \times 10^6$ axons
  – ~60%: primary motor c.
  – ~20%: premotor cortex
  – ~20%: somatosensory c.

• **Target**
  – Grey matter of the spinal cord on the contralateral side
    \[ \alpha \text{-motoneurones} \]
    – Direct – monosynaptic
    – Indirect – polysynaptic
    \[ \gamma \text{-motoneurones} \]
    – polysynaptic
Damage affecting the somatomotor areas (1)

• „Stroke” – ALWAYS affects the CONTRALATERAL regions of the body

• **Isolated damage of the Betz cells** (Experimentally only)
  – Does not result in paralysis, and does not induce alterations in the muscle tone
  – Inability to perform precise movements with the hands and fingers
Damage affecting the somatomotor areas (2)

• **Isolated damage of the primary motor cortex** (Experimentally only)
  - „Isolated” pyramidal damage
  - **Hypotonia** – lack of the tonic stimulation originating from the motor cortex and affecting spinal motoneurones.
Damage affecting the somatomotor areas (3)

- Simultaneous damage of the motor cortex and deeper structures (basal ganglia!) – ”STROKE”
  
  - **Acute**: hypotonia, areflexia, paralysis
  
  - **Chronic**: hypertonia, hyperreflexia, paralysis
Upper motoneurone syndrome

- "Upper motoneurone syndrome"
  - Damage of the descending motor pathways (anywhere; i.e. capsula interna)
  - Instantaneous flaccid paralysis and areflexia that is primarily affecting the limbs
  - Function of the trunk muscles is usually maintained:
    - Integrity of the brain stem
    - Bilateral innervation of muscles near the midline (bilateral projection of the corticospinal tract)
Upper motoneurone syndrome

• "Upper motoneurone syndrome"
  – After a couple of days, reactivation of the spinal local reflex circuits occur, markedly altering the general picture of the syndrome
  – Spasticity(hypertonia), hyper- and hyporeflexia, paralysis
  – Occurrence of the Babinski’s sign
Lower motoneurone syndrome

• „Lower motoneurone syndrome”
  – Only some of the muscles are affected (or even one)
  – Hypotonia, flaccid paralysis
  – Hypo- and areflexia
  – No “abnormal” reflexes
  – Marked and severe atrophy of the affected muscles